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Type 1 Cryoglobulinemia Secondary to Primary Plasma Cell Leukemia

Primary plasma cell leukemia (pPCL) is a rare and aggressive form of plasma cell dyscrasia that comprises 1% of the plasma cell dyscrasias. Even more rare is its association with type 1 cryoglobulinemia which management can be challenging. A 63-year-old African American male with no past medical history presented with severe burning pain in his legs, necrotizing rash, constitutional symptoms and severe acute kidney injury (AKI). HIV, hepatitis B and C were negative. Serum electrophoresis revealed an M-spike of 5.67, IgG of 7000, free lambda of 152.10 with a K/L of 0 and cryoglobulin of 81%. CT revealed extensive lytic lesions throughout the spine. Skin biopsy revealed thrombotic vasculopathy. Bone marrow biopsy revealed 80% dysplastic plasma cells. Flow cytometry of peripheral blood showed dysplastic plasma cells with high expression of CD38 and CD138 but negative for CD19, CD20, CD56 and CD117. FISH revealed t(11;14)(q13;q32) resulting in IGH/CCND1 gene rearrangement. Type 1 cryoglobulinemia secondary to IgG/lambda primary plasma cell leukemia was diagnosed. Patient's pain worsened and skin lesions increased in size and new eruptions appeared in other parts of the body, so plasmapheresis was initiated. After six plasmapheresis cycles, skin lesions stabilized, no new lesions appeared, pain regimen was weaned and AKI resolved. Cryoglobulin level became undetectable after four plasmapheresis cycles. VRD (bortezomib, lenalidomide, dexamethasone) was started during the hospitalization. No evidence of tumor lysis syndrome was noted. Patient was successfully discharged 3 weeks after presentation. Three months later, patient achieved a very good response. This case illustrates the rapidly progressive nature of type 1 cryoglobulinemia in patients with pPCL. Thus, recognition of severe organ dysfunction should raise consideration for plasmapheresis. In a case series by Payet et al., patients treated promptly with plasmapheresis achieve disease stabilization in 90% of the cases and those who had it delayed eventually required dialysis or skin surgery.